EDITORIAL

Pregnancy and the various forms of the Fontan circulation

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The Fontan operation is performed on those with highly complex congenital heart disease and has improved the survival and quality of life of such patients. Inevitably there are late complications secondary to extensive surgery and long-term survival estimates are about 70% at 10 years and 60–63% at 15 years. Pregnancy therefore requires careful prior consideration. However, the opinion that that pregnancy is not advisable for the Fontan patient is somewhat contentious and maybe an oversimplification of what is a highly emotive and complex issue.

Professor Jane Somerville once said to me in jest that the specialty of grown-up congenital heart disease (GUCH) would not be needed if it were not for the Fontan operation! Although this rather understates the case for many of the other congenital heart disorders which require long-term follow-up, it is a sentiment with which specialists in the field identify. The adult Fontan patient is plagued by a plethora of complications, often intractable and difficult to treat, and moreover their long-term survival is significantly reduced. It may come as a surprise to some, therefore, that Drenthen *et al* are reporting on pregnancy outcome in patients with a Fontan circulation[1]. They provide an excellent summary of their own and others' experience, but their final conclusion ''that pregnancy is inadvisable for the Fontan patient'' is somewhat contentious and in my opinion an oversimplification of what is a highly emotive and complex issue.

It is important to understand that the Fontan operation is performed on those with the most highly complex congenital heart disease, when a bi-ventricular repair is not possible. It was initially performed for those born with tricuspid atresia and sub-pulmonary stenosis, in whom the natural history was dismal without surgical intervention (~90% dying by the end of the first year of life).[2] The procedure has subsequently been modified many times over (fig 1) and its application extended to a wide spectrum of complex lesions including complete atrioventricular septal defects, double inlet left ventricle, double outlet right ventricle and pulmonary atresia with intact septum. It has improved the survival and quality of life for those born with these highly complex lesions and represents one of the landmark operations in the history of paediatric cardiac surgery.[3] The Fontan circulation is unique in that the sub-pulmonary ventricle is bypassed and systemic venous return is diverted directly into the pulmonary arteries, with the aim of providing adequate pulmonary blood flow and cardiac output with minimal elevation of venous pressure. Because the systemic and pulmonary venous returns are now separated, cyanosis and volume loading on the ventricle is relieved. The central venous pressure becomes the driving force for pulmonary blood flow and cardiac output is derived from pulmonary venous return. An additional forward driving force for pulmonary blood flow comes from spontaneous respiration and negative intra-thoracic pressure.[4]

Sadly, intrinsic to the construction of the Fontan circulation is the inevitability of complications secondary to extensive surgical distortion of cardiac anatomy and physiology. ''The Fontan state imposes a gradual declining functional capacity and premature death after an initial period of often excellent palliation'', salutary words from the pioneering surgeon himself.[3] The litany of late complications includes atrial dysrhythmias (about 46% at 10-year follow-up), myocardial dysfunction and failure (grade II, about 71% at 10-year follow-up), thromboembolic events (about 32% at 10-year follow-up), right atriomegaly and hepatic dysfunction, and protein-losing enteropathy (about 13% at 10-year follow-up), and long-term survival estimates are about 70% at 10 years and 60–63% at 15 years (atriopulmonary Fontans).[3 5 6] Bearing in mind that the Fontan operation is usually performed at approximately 4 years of age, one can anticipate problems for the majority of adult survivors. For those with newer modifications of the Fontan, for example total cavopulmonary circulation, late outcome and survival is not yet known.

In light of this unique finely balanced physiology and the numerous late complications, pregnancy requires careful prior consideration. It must also be borne in mind that the haemodynamic changes in pregnancy are not insignificant for the abnormal heart (cardiac output and stroke volume double, myocardial oxygen consumption increases by 20%, heart rate increases by 15–20%). The overriding principle of pregnancy care prioritises maternal health over that of the unborn child and an assessment of maternal risk and likely outcome is important. Nonetheless, foetal outcome is clearly elemental in this assessment and the likelihood of a healthy live-born infant, in the context of increased maternal risk, requires equally judicious appraisal. Moreover, the likely long-term consequences of pregnancy on the Fontan heart need to be contemplated on a background of a limited long-term survival.[5]

Drenthen *et al* have reported on their own experience of 10 pregnancies in six women and have also summarised the experience of others

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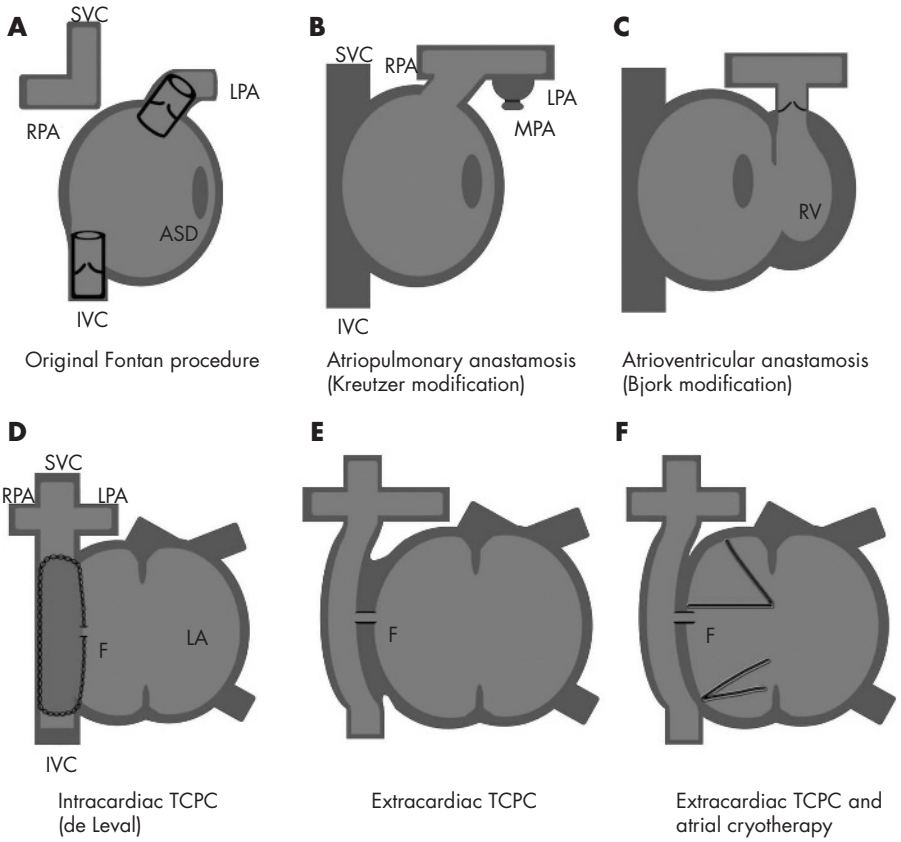

Figure 1 Various forms of the Fontan circulation. ASD, atrial septal defect; F, fenestration; IVC, inferior vena cava; LA, left atrium; LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava; TCPC, total cavopulmonary connection.

with respect to Fontan pregnancy outcome. There was no maternal mortality in their series, which is in keeping with the experience of others. However, there is morbidity, with atrial arrhythmias being most common. This is not surprising, as it is well known that almost half of the adult Fontan population develop atrial arrhythmias, even in the non-pregnant state.[5 7] A decline in functional status is also common. In Drenthen's series, two out of the four patients who completed pregnancy had a decline in NYHA status and from the combined total of 39 completed pregnancies reported in the literature, there was a decline in NYHA class in 16 and overt heart failure in four. Drenthen *et al* reported full recovery to baseline NYHA post-partum and this reflects my own experience in five completed Fontan pregnancies.

One might have expected an increase in thromboembolic complications in pregnancy, as both the Fontan state and pregnancy are pro-thrombotic milieu.[8 9] However, it is likely that those embarking on pregnancy will be fully anti-coagulated for this very reason and thus this complication is avoided. The corollary of full anticoagulation is that there is an increased risk of spontaneous abortion, irrespective of whether heparin or warfarin is used.[10] Drenthen *et al* reported a 50% spontaneous abortion rate, which is much higher than the normal population rate of 10–15%; however, they do not mention if their patients were anti-coagulated. This was also the experience of Canobbio *et al*, who reported on the largest Fontan pregnancy experience to date, of 33 pregnancies, where there were 15 live births and 13 spontaneous abortions.[11]

If pregnancy is sustained beyond 14 weeks, foetal outcome is usually good. Drenthen *et al* concluded that premature labour is higher than expected (1.25% for normal gestations), with overall 15 cases of the 39 completed pregnancies delivering before the 37th week and this clearly has an impact on neonatal outcome and infant mortality. Conversely, Canobbio *et al* reported a median gestational age of 38 weeks, with only one preterm labour, which is in keeping with my own data, with only one of the five completed pregnancies delivering before the 37th week.

Each Fontan patient must be individually counselled and advised of the risks and likely maternal and foetal outcome, taking account of functional status, ventricular function, arrhythmia burden, co-morbidity and objective exercise data where possible.[12] However, most Fontan women will more often than not still embark on pregnancy even when informed of the risks to their own health, the high spontaneous abortion rate, the increased risk of premature delivery, and their reduced longevity. The paper of Drenthen *et al* highlights this, reporting that 27 of the 35 childless Fontan responders were still contemplating pregnancy, including nine who had been advised against it by their physicians. Cardiology and obstetric services must therefore prepare to provide pregnancy care for these highly complex patients, which demands multidisciplinary teamwork and good communication between several specialists, all of whom should have expertise in managing such patients.[13]

The rationale and conclusions of Drenthen *et al* ''that pregnancy in the Fontan is inadvisable'' is contentious. It draws its conclusion from what is a very limited evidence base and it oversimplifies this emotive and complex issue. Although there is a risk of maternal morbidity, most complications are treatable and not insurmountable. Pre-conceptual counselling and careful individual risk assessment should ensure maternal mortality is a rarity. Foetal outcome is usually good when pregnancy is maintained beyond 14 weeks and although the rate of preterm labour may be increased, neonatal outcome is very much dependent on the degree of prematurity and

neonatal intensive care back-up. Clearly the psychological consequences of miscarriage are not inconsiderable, but they have to be weighed against the psychological impact of being told that one will be childless, and in my experience most GUCH mothers consider the latter more psychologically and emotionally distressing.

It is mainly when I contemplate the future for the Fontan mother and that of her child, that I have some disquiet. Specialists in the field are well aware that the third and fourth decades of life are likely to be beset by complications and functional deterioration. Nevertheless, Fontan patients, like many GUCH patients, have a different concept of the future and have often never thought ''old-age'' would be their reality.[14] They have lived daily with chronic disease and are constantly reminded of their own mortality, which ultimately gives them a different outlook on life. It is important therefore not to impose one's own ideas of adulthood or parenthood onto a person with such completely different life circumstances. With respect to the future of the child, it is for the prospective parents to bear this responsibility and plan the upbringing of their child in the context of their health, environment, and limitations just like anyone else. Our role as cardiologists is to assess risk and predict outcome as best we can and impart this knowledge, so that the patient is empowered to make their own choice, because ultimately it is they who have to live with it. It may challenge my doctrine that I always know what is best for my patient, but my role is not to play God but to provide medical care whatever their decision.

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